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Recurrent anti-GQ1b IgG antibody syndrome showing different phenotypes in different periods

Anti-GQ1b IgG antibody is often found in the sera of patients with Miller Fisher syndrome, Bickerstaff's brain stem encephalitis, Guillain–Barré syndrome with ophthalmoplegia, acute ophthalmoparesis without ataxia, and occasionally, in isolated internal ophthalmoplegia and chronic ophthalmoplegia.[1–3] These conditions may be designated as anti-GQ1b IgG antibody syndrome.[1] We report a patient who showed three different phenotypes of the anti-GQ1b IgG antibody syndrome at different periods.

Case report

The patient was a 19 year old woman. At age 10, she visited a neurologist because of diplopia and an unsteady gait two weeks after a respiratory tract infection. Neurological examination showed ophthalmoplegia, dilated pupils with sluggish pupillary responses, areflexia, and cerebellar ataxia. Laboratory findings including nerve conduction studies were normal except for a slight increase in CSF protein (40 mg/dl) without pleocytosis. Within three months, her condition gradually improved and she was discharged without a neurological deficit except for persistent areflexia.

At age 17, she noticed mild diplopia, which gradually got worse. She visited our hospital with the complaint of slowly progressive diplopia. Although ocular movements were not restricted, her pupils were dilated bilaterally with reaction to light. She showed areflexia and slight ataxia without pathological reflexes. Blood count, blood chemistry, and brain magnetic resonance imaging (MRI) were normal. Nerve conduction studies were normal except for absence of the F wave.

At age 19, a week after developing fever of unknown origin, the diplopia suddenly progressed. The next morning, her gait became unsteady and she was admitted to hospital. On admission (day 1), she had complete ophthalmoplegia without oculocephalic reflexes. The pupils were markedly dilated without reaction to light. Her speech was slurred. Her extremities were slightly weak. She could not sit on the bed by herself because of severe unsteadiness. Her deep tendon reflexes were absent, with no pathological reflexes. The CSF was normal, including the protein level. Nerve conduction studies were within the normal range except for absence of the F wave. An enzyme linked immunosorbent assay showed that serum IgG reacted strongly with GQ1b (titre, 1:3200) and GT1a (titre, 1:3200), but not with other gangliosides.

She was given intravenous immunoglobulin (IVIg) on days 2–6. On day 5, she developed disturbed consciousness, Cheyne–Stokes respiration, and extensor plantar responses. Intratracheal intubation was required for ventilatory failure. Electroencephalography (EEG) showed diffuse theta activity. Brain MRI was normal.

From day 11, her illness gradually improved. Serum anti-GQ1b IgG and anti-GT1a antibody titres decreased below the cut off level by day 55. The F wave on nerve conduction studies became normal by day 78. Fifteen weeks after the onset, she had almost recovered except for areflexia and slight restriction of ocular abduction of the both eyes.

Comment

Our patient showed three different conditions of the illness at three different periods between the ages 10 and 19: first, acute onset of ophthalmoplegia, ataxia, and areflexia at age 10, which is a typical presentation of the Miller Fisher syndrome; second, chronic progressive diplopia associated with internal ophthalmoplegia from age 17; and third, acute onset of complete ophthalmoplegia, ataxia, marked drowsiness, and respiratory paralysis with extensor plantar responses and EEG abnormalities at age 19.

We diagnosed the third episode as Bickerstaff's brain stem encephalitis, because she showed transient central nervous system involvement (drowsiness, respiratory disturbance, positive plantar responses, and EEG abnormalities) in addition to the triad of the Miller Fisher syndrome. High anti-GQ1b and anti-GT1a IgG antibody titres at the time of the most recent illness and their decrease following recovery supported this diagnosis.

There are clinical similarities between Miller Fisher syndrome and Bickerstaff's brain stem encephalitis, and these conditions have been considered as consecutive spectra of the same disease. Miller Fisher syndrome is usually a monophasic illness, but, on rare occasions, it may recur after a long asymptomatic interval.[4 5] It has been reported that clinical features of recurrent Miller Fisher syndrome are constant from episode to episode.[4 5] This is in contrast with recurrent Guillain–Barré syndrome, which shows considerable variety in the distribution and severity of weakness between each episode.[4] This is the first report of Bickerstaff's brain stem encephalitis as a recurrent episode of the Miller Fisher syndrome.

In the second phase of chronic progressive diplopia, our patient showed abnormalities of the pupils with slight ataxia and absence of the F wave in nerve conduction studies. As external ocular movement was not restricted, progressive diplopia might reflect pupillary abnormalities; diplopia has been discussed in isolated internal ophthalmoplegia without external ophthalmoplegia associated with anti-GQ1b IgG antibody.[2] In addition, chronic external ophthalmoplegia has been found with raised serum anti-GQ1b IgG antibody.[3] These findings suggest that chronic internal ophthalmoplegia may be associated with anti-GQ1b IgG antibody, although we could not examine this antibody during that period.

It is unique in our patient that three different phenotypes of the anti-GQ1b IgG antibody syndrome appeared at different times. There has up to now been no report in which different anti-GQ1b IgG antibody syndromes have recurred at different times in a single patient. Our case indicates that Miller Fisher syndrome, Bickerstaff's brain stem encephalitis, and chronic internal ophthalmoplegia form part of the spectrum of the anti-GQ1b IgG syndrome, although the mechanism of the variability in clinical phenotypes of the anti-GQ1b IgG syndrome remains unknown.

In conclusion, our case indicates that different phenotypes of the anti-GQ1b IgG antibody syndrome can occur at different times in the same patient, showing that this syndrome may be a distinct entity with a wide clinical spectrum on a unique immunological background.

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Sydenham's chorea may be a risk factor for drug induced parkinsonism

Sydenham's chorea, the most common cause of acquired chorea in childhood, is a delayed complication of group A β-haemolytic streptococcal infection.[1] It is thought to be caused by antibodies induced by streptococci which cross react with basal ganglia antigens.[2] Despite the decrease in Sydenham's chorea in developed countries, there is a renewed interest in this condition because of the hypothesis that a similar mechanism may play a role in the pathogenesis of a subset of patients with tics and other neuropsychiatric disorders.[3]

The treatment of Sydenham's chorea is based on the combination of penicillin and antichoreic drugs (valproic acid and/or dopamine antagonists). At the movement disorders clinic of the Federal University of Minas Gerais (MDC-UFMG), located in an area where Sydenham's chorea remains endemic, we have been struck by the occurrence of drug induced parkinsonism among patients with Sydenham's chorea. We therefore